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Thoracic endometriosis presenting as hemopneumothorax

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Abstract

Thoracic endometriosis is very rare. Usually, the thorax is the most frequent affected site outside the pelvis. Common symptoms include chest pain, dyspnea, and hemoptysis. Common manifestations include pneumothorax, hemothorax, and pulmonary or pleural nodules. In addition, symptoms and manifestations can be "catamenial" happening a few days after menstruation onset. This disease can be debilitating, causing a significant impact on the quality of life of young women. We present a case of a young female who was referred to our hospital with recurrent right-sided pleural effusions and pneumothoraces. Pleural fluid drainage was consistent with hemothorax. Transvaginal ultrasound showed mild intraperitoneal fluid in the Cul-de-Sac. Due to concerns for thoracic endometriosis, video-assisted thoracoscopic surgery was performed confirming the diagnosis by pathology. Therapeutic pleurectomy with diaphragmatic repair and pleurodesis was performed. The patient was started on medroxyprogesterone acetate injections two weeks after with great clinical response.

Introduction

Thoracic endometriosis syndrome (TES), characterized by the presence of ectopic uterine endometrial tissue in the thorax, is very rare [1]. The main manifestations include pneumothorax, hemothorax, and pulmonary and pleural nodules [2]. Symptoms include chest pain, dyspnea, hemoptysis, and abdominal pain. Clinical and radiologic manifestations are termed "catamenial" if related to the menstrual cycle and onset of menses; however, this relation is not always present [3]. Usually, the disease is manifested predominantly in the right thorax [3,4]. Treatment can be challenging, and recurrence is common [5,6]. In refractory cases to tube thoracostomy drainage and hormonal suppressive therapy, video-assisted thoracoscopic surgery (VATS) with resection, pleurodesis and diaphragmatic repair might be curative [5-8]. A multidisciplinary approach to TES including coordination between thoracic surgery and gynecology might improve the quality of life and outcomes of affected women [9]. We report a case of TES presenting with recurrent hemopneumothoraces requiring multiple pleural drainages, VATS, and hormonal suppressive therapy.

Case Report

The patient in this report was a 33-year-old female G2P1A1 with a history of dilation and curettage a few years earlier, and a history of previous intrauterine copper contraception device (IUD) placement and retrieval. She presented to our facility with worsening shortness of breath and right-sided chest pain for four

months duration. She reported worsening of her symptoms a few days after menses. She reported occasional heavy menses and dysmenorrhea. Four months before presentation, she was treated for possible pneumonia and had a thoracentesis performed in an outside facility in a different state with the pleural fluid looking bloody per patient. She was also informed she had a small "air leak" on her chest X-ray. She left against advice at the time, before the completion of her care.

On presentation, she was hemodynamically stable. She had decreased air entry with dullness to percussion over the right hemithorax. The hemoglobin level was low at 9.2 g/dL and a hematocrit of 29.8%. A chest X-ray revealed a moderate size pleural effusion (Figure 1A). A CT-scan of the chest confirmed the presence of a moderate size non loculated, non-septated pleural effusion with compressive atelectasis of the right lower lung lobe (Figure 1 B,C). However, there was no evidence of pleural, parenchymal or diaphragmatic involvement suggested by imaging.

After discussing the treatment options, the patient elected to undergo a thoracentesis. A transthoracic bedside chest ultrasound revealed a moderate size, non-loculated, non-septated, free-flowing pleural effusion with a simple fluid density as well as compressive atelectasis of the lung (Figure 2A). The ultrasound failed to show any involvement of the pleura, diaphragm, or lung parenchyma. An ultrasound-guided thoracentesis was performed, and 1.2 liters of bloody fluid was recovered and sent for analysis (Figure 2B). The pleural fluid analysis returned exudative based on Light's criteria for lactate dehydrogenase (LDH) and protein criteria. It was also consistent with hemothorax as the pleural fluid hematocrit was 16%, more than half of the serum hematocrit. Pleural eosinophils were elevated as well constituting 26% of total leukocytes. Gram stain and culture as well as cytology, cell block, and flow cytometry were negative for infection or malignancy.

A few days later, a chest X-ray showed a new moderate-size pneumothorax and residual right-sided small pleural effusion. (Figure 3A). A repeat CT-scan of the chest showed a hydropneumothorax (Figure 3 B,C) as well; still without any radiologic evidence of pleural, parenchymal, or diaphragmatic involvement by the disease. A transvaginal ultrasound showed normal uterus and ovaries with mild peritoneal fluid in the Cul-De-Sac (rectouterine pouch) suggestive of a small volume hemoperitoneum that can be encountered with endometriosis (Figure 3D). The patient remained in no distress and breathing room air. After discussion with thoracic surgery, the plan was to undergo VATS for diagnosis and therapy as there was high suspicion of TES.

Intraoperatively, bloody pleural fluid was encountered and suctioned during chest entry. Fiberoptic inspection of the chest cavity showed large dark implants on the parietal pleura at the base of the diaphragm, which was excised and sent for pathology. In addition, large diaphragmatic fenestrations were noted allowing the visualization of the liver through the chest. These defects were sutured and repaired. Complete parietal pleurectomy was performed, followed by mechanical pleurodesis and chemical pleurodesis with doxycycline. Post-operative imaging revealed resolution of the hemopneumothorax, and the patient had successful pleurodesis. Pathology from the diaphragmatic implants and pleura showed endometrial tissue consistent with endometriosis (Figure 4).

The patient followed up with gynecology and pulmonary after discharge and was started on monthly injections of

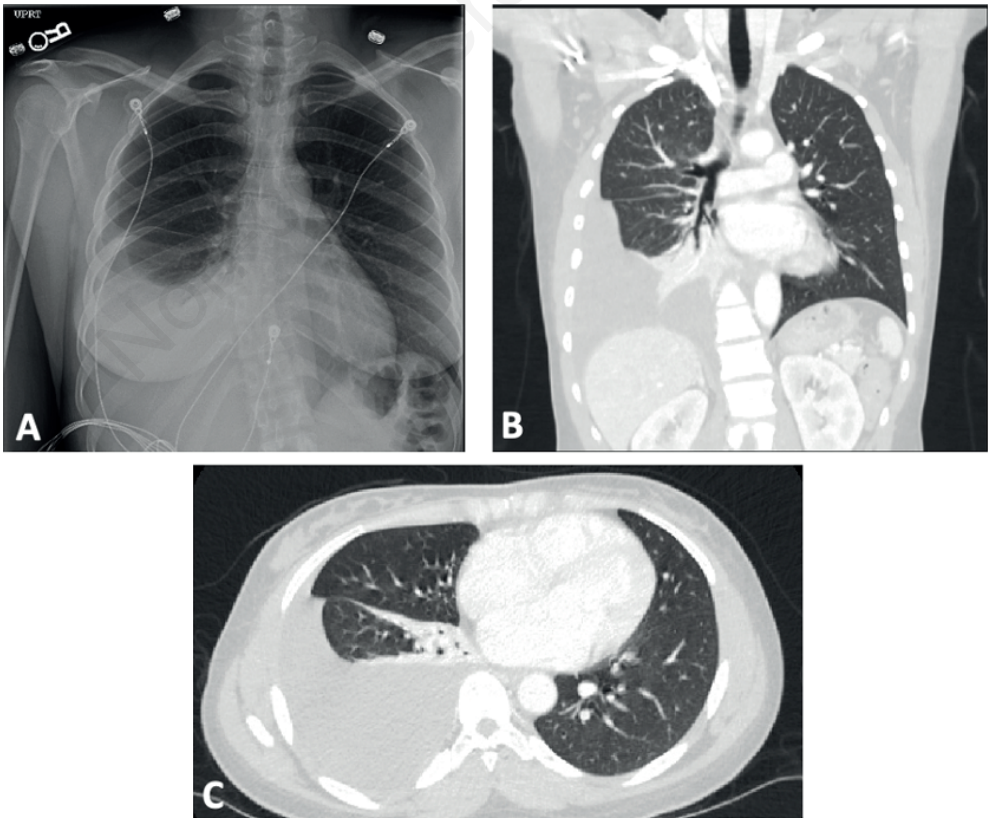

Figure 1. A) Chest X-ray showing right-sided pleural effusion. B,C) CT-scan of the chest showing moderate size pleural effusion with compressive atelectasis of the right lower lobe.

medroxyprogesterone acetate to control her disease burden and decrease thoracic recurrence. At 1-year follow-up after surgery, the patient was asymptomatic and reported no dyspnea or chest. A 1-year follow-up chest X-ray showed expected post-operative changes, but no recurrence of the pleural effusion or any visible pneumothorax.

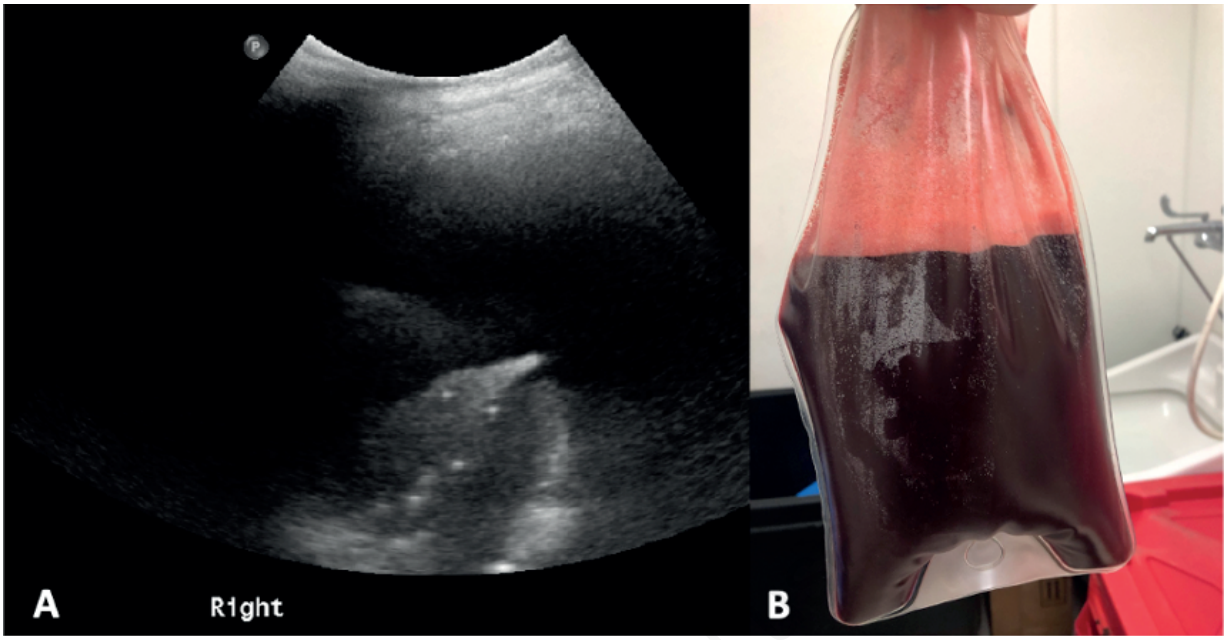

Figure 2. A) Transthoracic ultrasound showing right-sided, moderate size, non-loculated pleural effusion. B) Bloody "chocolate" colored recovered pleural fluid.

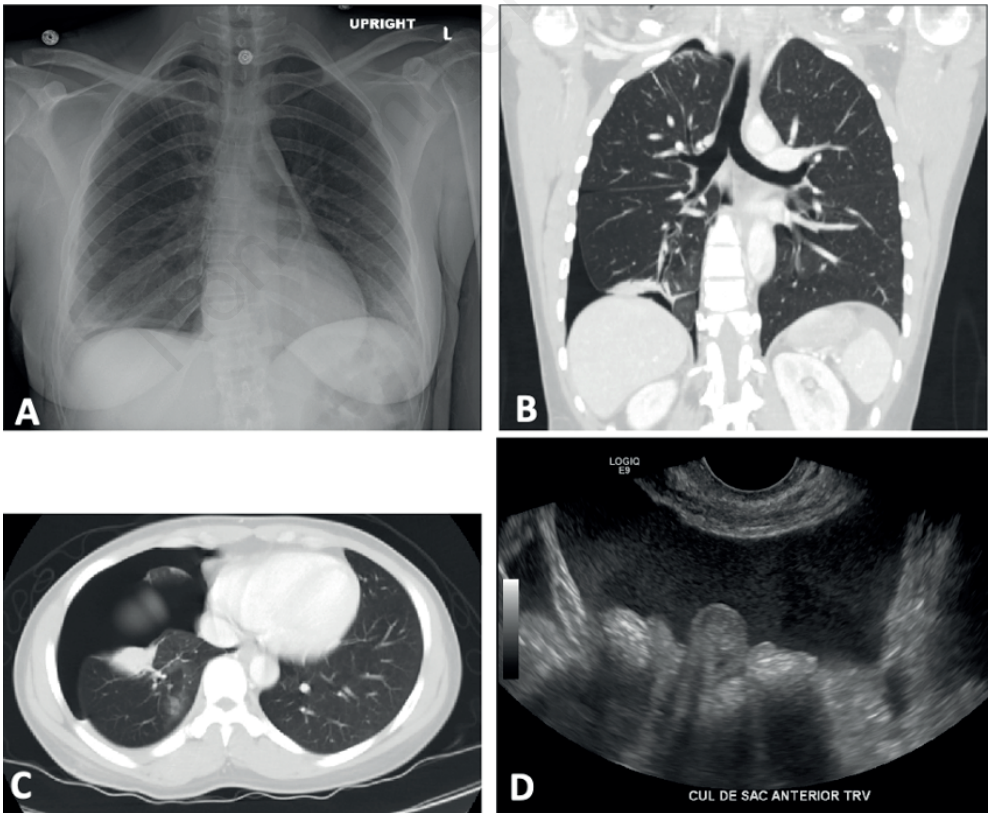

Figure 3. A) Chest X-ray showing a pneumothorax with small right pleural effusion. B,C) CT-scan of the chest showing moderate size right pneumothorax with residual small right pleural effusion. D) Transvaginal ultrasound showing mild peritoneal fluid.

Discussion

TES is very rare and occurs in around 1% of patients with pelvic endometriosis [10]. In childbearing-aged women presenting with cyclical chest pain, pneumothorax, hemothorax, or hemoptysis, TES should be high on the differential [11]. In patients with a history of pelvic surgery and endometriosis, there is a higher prevalence of TES [12]. This was the case in our patient who previously had dilation and curettage as well as a previous placement of a copper IUD.

Manifestations of TES are predominantly right-sided; with chest pain and dyspnea being the main symptoms and pneumothorax being the most commonly observed pathology [1-4]. Hemothorax and hemopneumothorax are less frequently encountered and are seen in around 10% to 15% of patients with TES [2,4]. Hemothorax is usually seen in patients with increased pleural disease burden [4]. The patient in this report had recurrent hemopneumothorax however her records from the outside facility could not be obtained to confirm. Her disease was mainly right-sided and included both pneumothorax and hemothorax. The CT-scan of the chest after the thoracentesis did not reveal any pulmonary nodules, parenchymal disease, or diaphragmatic lesions. However, VATS confirmed the presence of significant parietal pleural disease and diaphragmatic implants and fenestrations.

A multidisciplinary approach to her disease including management by thoracic surgery and gynecology has been shown to improve outcomes and decrease recurrence [8,9]. For recurrent disease, VATS is preferred along with occasional concomitant laparoscopy to evaluate abdominal tumor burden [1,2,7,9]. Due to the non-significant transvaginal ultrasound, the patient underwent only VATS without laparoscopy with plans to manage the abdominal disease with hormonal therapy. Diaphragmatic defects and fenestrations are seen in around 25% of patients with TES and might complicate the course of the disease [5]. Pleural fluid cytology yield is low at 10%, while surgical pleural cytology yield is around high at 80% [1,12]. Therefore, if clinical suspicion for TES is high in the proper clinical setting, the need for VATS for diagnosis and therapy becomes essential. Interestingly, multiple imaging modalities used for our patient including X-ray, CT-scan and ultrasonography failed to show involvement of the parietal pleura, diaphragm and parenchyma. The involvement was however encountered and confirmed during VATS. Therefore, the true extent of the disease in the thoracic cavity might be underestimated with imaging and should diminish the importance of surgery.

It is recommended to treat for at least 6 months with hormonal suppressive therapy to prevent recurrence of TES, and progression of disease prior to or after surgery [2,7,13]. The patient in this report tolerated monthly injections of medroxyprogesterone in the gynecology clinic with no side effects for at least one year with significant improvement in her quality of life, and symptoms.

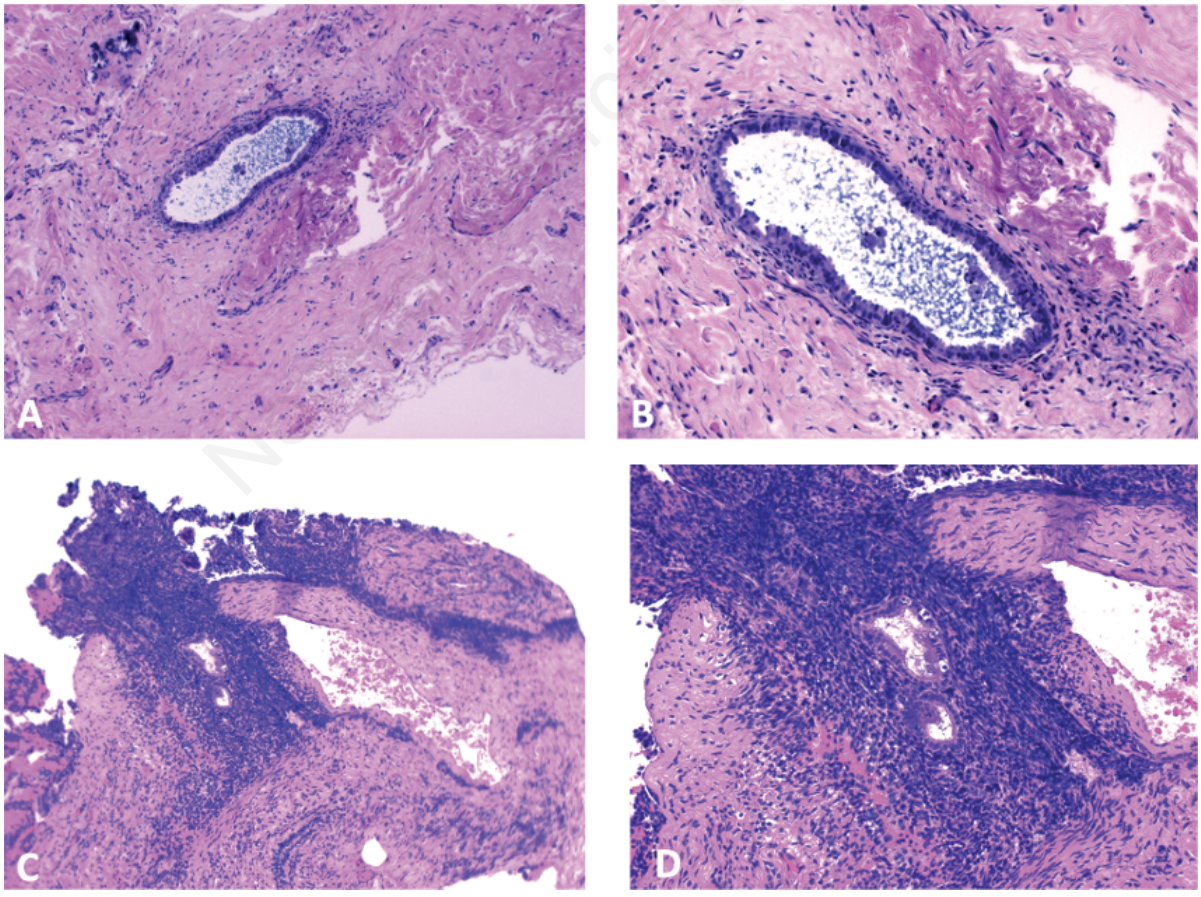

Figure 4. A,B) Hematoxylin-and-eosin-stained slides showing ectopic Müllerian type gland within the eosinophilic fibrous tissue of the parietal pleura; the gland has simple columnar epithelium and a thin rim of more basophilic endometrial stroma; there are several hemosiderin-laden macrophages within the gland, which is characteristic of endometriosis. C,D) Hematoxylin-and-eosin-stained slides showing two ectopic Müllerian type glands in a portion of the diaphragm; the glands are in a background of basophilic endometrial stroma.

Conclusions

TES is a rare cause of pneumothorax, hemothorax, chest pain, or hemoptysis. It should be suspected in women of childbearing age, especially with a prior history of pelvic surgery. Most common symptoms include right-sided chest pain and dyspnea. Usually, the thoracic disease is right-sided with pneumothorax being more common than hemothorax. Treatment is multidisciplinary involving chest tube drainage if needed, hormonal suppressive therapy, a gynecologic evaluation, and ultimately thoracic surgery for refractory disease. A team approach decreases the chances of recurrence and improves the patient's quality of life.

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